

to detect cystic macular lesions, while clinical funduscopy examination did not.

Finally, we are fully aware of the genotypic, phenotypic, and electrophysiological differences between retinitis pigmentosa and the enhanced S-cone syndrome. None of the patients who were included in our study¹ showed any clinical or ERG features suggestive of the enhanced S-cone syndrome or the Goldmann-Favre syndrome variant.

Based on the sentiments expressed above, we do not accept the suggestion by Ganesh and colleagues that it would be prudent to perform FFA in "all" retinal dystrophy patients with a cystic maculopathy. Optical coherence tomographic testing would be preferable for screening based on its superior sensitivity for detecting cystoid macular changes compared to FFA as well as for patients' comfort and convenience.

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Ocular Adnexal IgG4-Related Lymphoplasmacytic Infiltrative Disorder and Graves Ophthalmopathy

We read with interest the article by Bijlsma and Kalmann.¹ The authors described a patient who had both idiopathic orbital inflammation (IOI) and Graves ophthalmopathy (GO). This was unexpected because each has distinct clinical character-

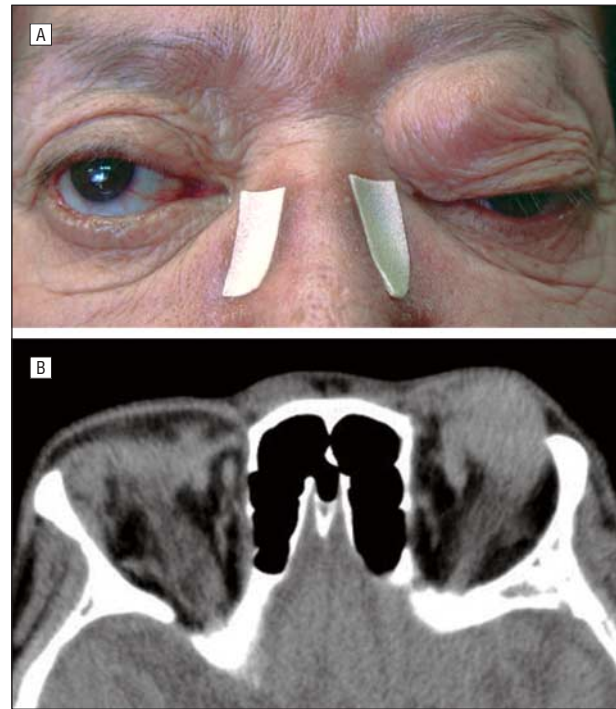


Figure 1. A, External photograph of patient showing exophthalmos, hypertropia, exotropia, and left upper eyelid swelling. B, Computed tomography shows that the lesions have infiltrated into the anterior superior orbits.

istics. They proposed a yet-undetermined common pathogenesis for these disorders.¹ We present clinical data obtained from a patient that can explain a relationship between these distinct clinical entities.

A 64-year-old woman with a long history of GO associated with hyperthyroidism was referred to our hospital for treatment of a left upper eyelid tumor (**Figure 1**) in April 2010. Palpation and computed tomography revealed anterior superior orbital infiltrative lesions in both eyes (Figure 1). Results of serological testing were positive for thyroid stimulating hormone receptor antibody, thyroid stimulating antibody, and antithyroid peroxidase antibody. The patient also had an elevated serum IgG4 level (1000 mg/dL; reference range, 4.8-105 mg/dL; to convert to grams per liter, multiply by 0.01) and IgE (1615 µg/L; reference range, <170 µg/L; to convert to milligrams per liter, multiply by 0.001). Histologic analysis of a biopsy specimen revealed sclerosing inflammation with IgG4-positive plasma cells (**Figure 2**). The histological and serological results met the criteria for ocular adnexal IgG4-related lymphoplasmacytic infiltrative disorder.² The orbital lesions decreased in size with oral prednisolone (30 mg), with slow tapering.

Patients with ocular adnexal IgG4-related lymphoplasmacytic infiltrative disorder have clinicopathologic characteristics that are unique in patients with IOI; this diagnosis may be associated with the immunologic imbalances by activated helper T cells type.² Graves disease is considered an autoantibody-mediated, T helper-dominant disease.^{3,4} Thus, we suggest that ocular adnexal IgG4-related lymphoplasmacytic infiltrative disorder and GO can be encountered simultaneously not as an inci-

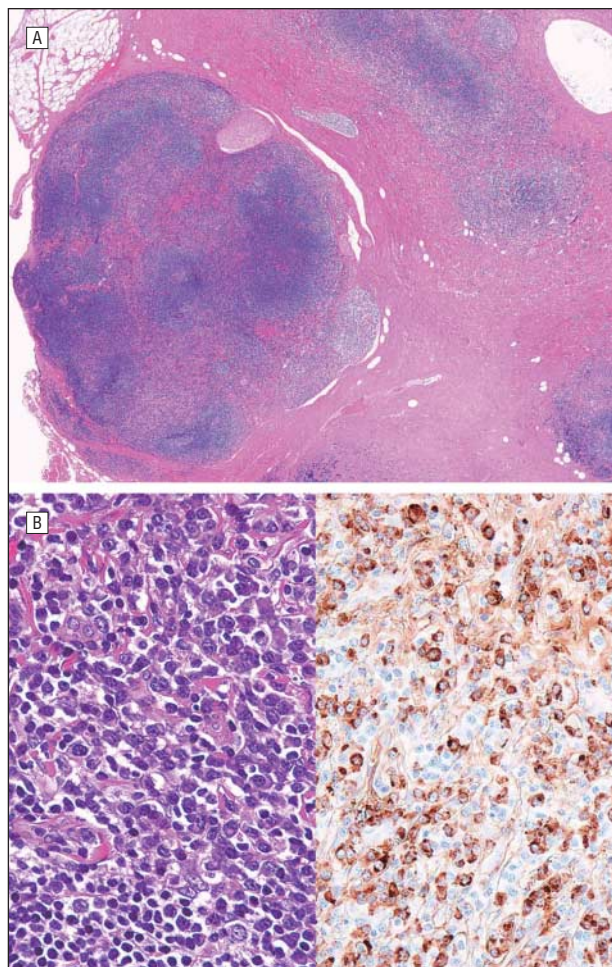


Figure 2. Histology and immunohistochemistry of an ocular adnexal lesions. A, Low magnification shows sclerosing inflammation with a reactive lymphoid infiltrate (hematoxylin-eosin, original magnification $\times 40$). B, High magnification show lymphoplasmacytic infiltrations and immunostaining for IgG4 reveals IgG4-positive plasma cells (IgG4 to IgG ratio, 80%) in the reactive lymphoid lesions (hematoxylin-eosin and immunoperoxidase, original magnification $\times 400$).

dental finding but because they are related to a similar systemic condition.

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ter the procedures and possible outcomes were explained to them.

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In reply

We would like to thank Kubota and colleagues¹ for sharing their case that may help explain the link between GO and IOI. In response, we performed histology on IgG4 on available specimens of our cases. Samples were available for case 1 and 4. Only case 4, who had developed unilateral IOI at 22 years of age and GO at 27 years, showed greater than 30% IgG4-positive plasma cells. We did not have the opportunity to determine IgG4 serum levels. Typically, IgG4-related ocular adnexal infiltration presents at a later age and is more often bilateral.¹ We agree that T helper cell imbalance may explain the co-occurrence of IOI and GO, but because of negative IgG4 findings in another case, other common pathogenetic causes may also exist.

The increasing number of reports of IgG4 related orbital inflammation raises the question if this condition should be regarded as a specific IOI entity like sarcoidosis and Wegener granulomatosis. However, the question has still to be answered whether IgG4 is only a marker of 1 or multiple underlying diseases or that IgG4 is in itself an inciter of orbital inflammation.

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Norrie Disease vs Familial Exudative Vitreoretinopathy

We thank Kiernan and colleagues¹ for their interest in and comments on our article.² We have read their related article³ and would respectfully like to argue that their patient has stage 2 familial exudative vitreoretinopathy (FEVR), not Norrie dis-